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Case Report

Fibrous dysplasia of the paranasal sinuses: case report

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ABSTRACT

Fibrous dysplasia (FD) is a rare congenital non-malignant bone tumor that affects the skeletal system development and interferes with normal bone formation. It's non-hereditary and its exact cause remains unknown. The condition is characterized by fibro-osseous connective tissue gradually replacing normal bone and marrow leading to the formation of bone that's fragile and prone to fractures. It is not uncommon for the disease to affect craniofacial bones but the sinonasal cavity involvement remains extremely rare. We presented a case of an 80-years-old male patient who reported nasal obstruction and a long-term swelling in the upper jaw that dates back over 20 years resulting in facial deformities and unilateral proptosis from compression exerted on the eye by the tumor. The patient reported having taken drugs to treat symptoms especially nasal congestion. But his dyspnea persisted. Physical examination, computed tomography (CT) scan and MRI have been elaborated and discussed. And on last resort, surgery was recommended.

Keywords: Fibrous dysplasia, Paranasal sinuses, Optic nerve, Facial deformity

INTRODUCTION

Albright described fibrous dysplasia (FD) for the first time in 1937 as a condition where normal bone is progressively replaced by fibrous tissues resulting in bone weakness that makes it fragile and prone to fractures.[1]

It is a congenital, non-hereditary, chronic disorder that affects the skeletal system. The disease shows no gender predilection as it affects equally both males and females. Most cases, about 75%, are reported in young individuals aged under 30 years.[1]

The disease occurs mainly among children as it begins in the childhood and continues throughout puberty and adolescence. It was believed that the disease goes dormant in early adult life, but recent research seems to support the idea that fibrous dysplasia may progress to adulthood. FD comprises 2% of all bone tumors and 7% of all benign bone neoplasms.[3,5]

FD can affect any bone in the body, but long bones such as femur, tibia and humerus are the most commonly affected. The craniofacial bone is also often involved especially maxilla, orbit, mandible and, much less frequently affected, the ethmoid bone. The disease, however, is rarely reported in paranasal sinuses and is usually subsequent to the extension of the disease from adjacent bones. Fibrous dysplasia remains asymptomatic until there is pain, or some adjacent structure vital function is compromised under the pressure exerted by the extending tumor.[3,5,6]

CASE REPORT

An 80-years-old man was referred to our hospital with bilateral chronic nasal obstruction, and a swelling in the

upper right maxillary that started more than 20 years ago. The lesion gradually and insidiously developed to reach the present size.

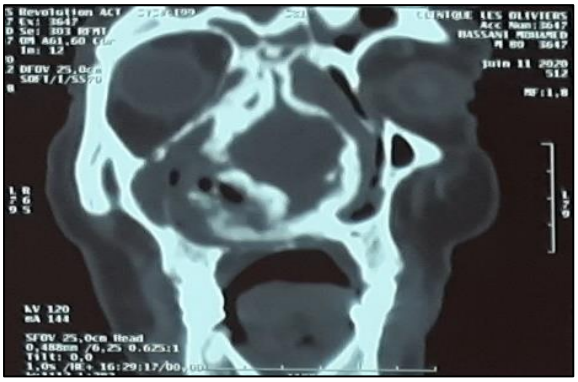

Figure 1: Computed tomography demonstrate the total obstruction of nasal airway.

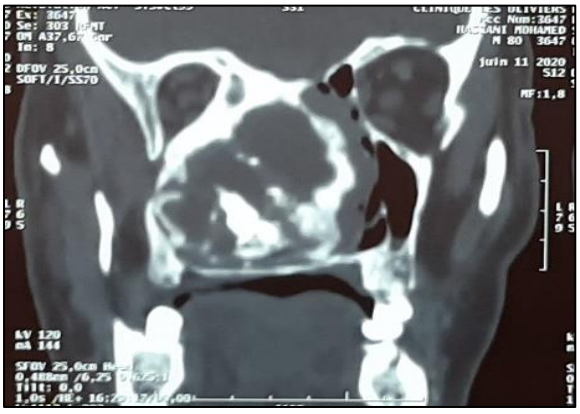

Figure 2: Computed tomography scan showed the disease expanding to right orbit.

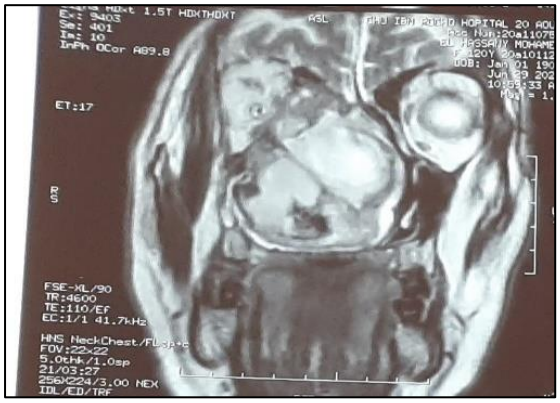

Figure 3: MRI scan (coronal slice) reveals the expansion of the FD to the right eyeball.

He was treated previously with topical corticosteroids and decongestants but failed to reduce his symptoms. No known family history was revealed by the patient. No traumatic history was present. On extraoral examination gross facial asymmetry was noticed on right side of the face, with deviation of nose to left side. We observed a mild proptosis of the right eye.

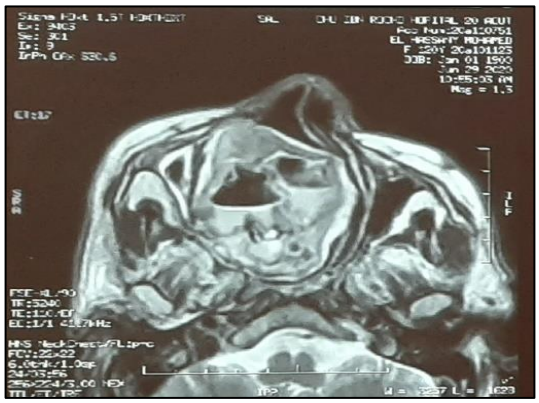

Figure 4: MRI scan showed the expansion to the whole nasal cavity.

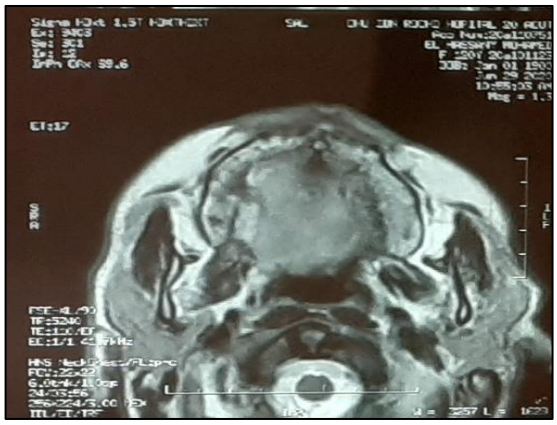

Figure 5: MRI scan (axial slice) demonstrate the expansion of the upper maxillary.

On the nasal endoscopic examination showed a left deviated septum. It's worth mentioning that the patient declined to undergo surgery.

The CT scans showed characteristic 'ground-glass' appearance of the base of skull, including the frontal sinus, sphenoid sinus, ethmoid complex, maxillary sinus and middle turbinate with poorly defined borders and normal mucosal covering, which was compatible with FD.

DISCUSSION

Patients with fibrous dysplasia commonly suffer from facial asymmetry and painless swelling of jaws. And in a few cases headache or bone pain, nasal discharge and obstruction were reported. The disease can also cause cranial nerves compression which leads to anosmia, hearing loss, facial paresis, visual impairment, exophthalmos, ptosis and eyelid edema.[5,6] The exact cause of fibrous dysplasia remains unknown. It is believed that the defect is due to a mutation in a gene

called GNAS1 that instructs bone formation and growth. The genetic mutation occurs in the early stages of fetal development and affects both sexes indiscriminately. The disorder results in abnormal fibro-osseous tissue instead of healthy normal bones. The disease can manifest itself at any age, but it usually appears during periods of bone growth which means childhood and adolescence.[7,8]

Under the microscope, FD is presented as a trabecular structure resulting from abnormal arrangement of bone tissue.[8] Biopsy is not always required unless there is an exacerbation of symptoms. Asymptomatic patients usually require a long-term follow-up.[7,8]

There are three types of fibrous dysplasia: monostotic (involving one single bone) is considered the most common as it represents 80% of all cases, polyostotic (involving multiple bones) accounts for nearly 20% and type 3 also known as McCune-Albright syndrome which involves the dissemination of pathological bone changes connected to other protuberances.[7] Malignant transformation is rare and represent only 0.5% and is usually observed in polyostotic cases.[8] Fibrous dysplasia presents a diagnostic challenge and can easily be mistaken for osteoma or ossifying fibroma. Thus, it is important to include these conditions in the list of differential diagnoses of the disease. Other diseases include, aneurysmal bone cyst, osteosarcoma, osteochondroma, cemento-osseous dysplasia, eosinophilic granuloma and deforming osteitis.[8,9]

A computerized tomography (CT) scan is the primary technique for diagnosis of fibrous dysplasia. The condition may be easily diagnosed even in an asymptomatic person. CT scans provide us with detailed images of the bones. The lesions are classified into three major types: pagetoid characterized by equal rate of the bone–fibrous matrix, sclerotic characterized by a high-density signal on CT and affects commonly facial and skull base bones and finally cystic pattern.[9]

Unlike CT scans, MRI allows us to differentiate the soft tissue character and visualize the intracranial growth of the tumor. MRI signal is hypointense on both sequences T1 and T2 with some heterogeneity observed on the latter.[9,10] In most cases fibrous dysplasia is asymptomatic and requires no treatment. The nature of the intervention is determined by the severity of the disease which, in turn, depends essentially on the location and the damage inflicted on neighboring organs. In an effort to relieve pain and prevent symptoms surgery might be indicated in spite of the controversy surrounding it. Surgery ranges from conservative excision to complete excision with the latter being always preferred and recommended.[10] Asymptomatic individuals with fibrous dysplasia do not need treatment. But patients presenting symptoms such as blocked sinus ostium leading to excruciating headaches and blocked sinonasal tract causing nasal obstruction and purulent discharges, would require endoscopic surgery to relieve pain and restore normal nasal breathing. Craniofacial fibrous dysplasia swellings may also cause acute facial disfigurement and complex ophthalmic manifestations such as visual impairment, oculomotor nerve dysfunction, proptosis and eyelid edema which would also require endoscopic surgery to decompress the optic nerve and prevent visual loss.[11]

One technique that is often performed is endonasal endoscopic tumor removal with its known benefits of minimally invasive surgery. As opposed to open surgery, this technique is associated with shorter hospital stay, increased and localized accuracy, less pain, faster recovery, less external scars and shorter recovery time. And as with any other type of tumor, fibrous dysplasia recurrences were reported, and patients should be monitored and examined periodically.[11,12]

Surgery is not always unavoidable since medications such as bisphosphonates can help prevent bone fractures and ease pain. It is believed that bisphosphonates can slow the disease's progression as they inhibit high bone turnover and are well tolerated in most patients, although some publications suggest that they are involved in patients developing osteonecrosis of the jaw. Zoledronic acid and pamidronate are also well known for their effects in controlling disease activity.[13,14]

CONCLUSION

Fibrous dysplasia is a fairly rare and benign pathology that often goes unnoticed without any treatment or medical intervention. But certain individuals may experience severe symptoms ranging from aesthetic discomfort to functional decline such as visual impairment. Surgery is usually recommended in the case of a significant facial disfigurement or to decompress the adjacent vital structure. Surgical treatment may also help ease the patient's pain. Needless to say, it's of high importance to remove as much damaged tissue as possible while preserving the patient's vital functions and organs.

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